clinical pediatrics

Index To Volume 11

January 1972—December 1972

Philadelphia Montreal

J. B. LIPPINCOTT COMPANY

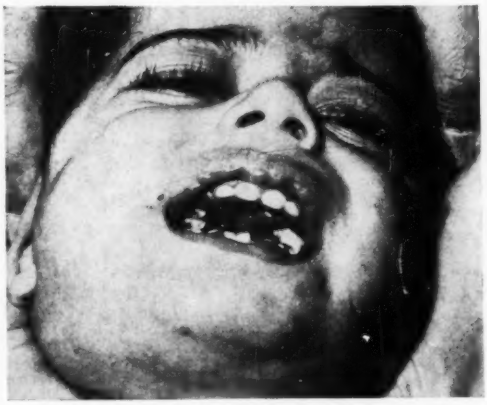

FIG. 1. A 16-month-old girl with malpositioned teeth and severe gingival swelling extending to the arch of the palate.

FIG. 2. A discrete yellowish-brown, scaly maculopapular rash covers the sternum and upper back.

Comments

This case of Letterer-Siwe disease had the following rather unusual and puzzling features: on initial examination, the patient had oral manifestations; the peripheral white blood cell and platelet counts were normal; the bone marrow picture was not characteristic; radiologic skeletal lesions were absent; and there was laboratory evidence of hepatic dysfunction.

In this disorder, oral manifestations, though uncommon, do occasionally occur. Leukopenia and thrombocytopenia may not be present early in the disease. In the initial stages, the bone marrow may not be involved, but will eventually be extensively replaced by proliferating histiocytes leading to myelophthisis and lowering of all cellular elements in peripheral blood.

When Letterer-Siwe disease is suspected clinically in a patient whose bone marrow is negative (such as our patient), the diagnosis is best confirmed by biopsy. There is no other specific laboratory approach. Typical localized bone destruction is frequent, especially in the calvarium. Our patient was not jaundiced, but her laboratory studies were consistent with subclinical hepatic dysfunction.

Clinical Pediatrics—Subject Index, 1972